

Fibrous Dysplasia of the Second Cervical Vertebra

A CASE REPORT*

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In patients who have polyostotic fibrous dysplasia, involvement of the vertebral column is rare; in patients who have the monostotic form of the disease, it is even rarer (Table I)^{3,4,9-12,15}. The series of Harris et al. included thirty-seven patients who had polyostotic lesions; three had in-

a monostotic spinal lesion: two lesions involved the lumbar spine; one, the thoracic spine; and three, the cervical spine^{6-8,13,14}. All of these lesions were identified after an injury had occurred; in two patients, neurological deficits had developed. We report the case of a patient who had an isolated lesion of the second cervical vertebra, which was identified incidentally when a whole-body bone scan was made for a lesion in the lower limb.

TABLE I

VERTEBRAL INVOLVEMENT BY FIBROUS DYSPLASIA

Series	Type of Fibrous Dysplasia	No. of Patients	No. of Patients Who Had Vertebral Involvement*
Stephenson et al., 1987	Monostotic	24	2†
	Polyostotic	19	2†
Dahlin and Unni, 1986	Monostotic	418	6
	Polyostotic	53	NA
Henry, 1969	Monostotic	50	0
Firat and Stutzman, 1968	Monostotic	15	0
	Polyostotic	9	2
Harris et al., 1962	Monostotic	13	1
	Polyostotic	37	3 (cervical)‡ 6 (lumbar)
Case reports			
Stirrat et al., 1989	Monostotic	1	1 (cervical)
Kahn and Rosenberg, 1988	Monostotic	1	1 (lumbar)
Troop and Herring, 1988	Monostotic	1	1 (lumbar)
Rosenblum et al., 1987	Monostotic	1	1 (thoracic)
Resnick and Lininger, 1984	Monostotic	1	1 (cervical)
Rosendahl-Jensen, 1956	Monostotic	1	1 (cervical)
Jaffe, 1946	Polyostotic	1	1

* NA = not available.

† Includes pelvic and spinal lesions.

‡ Numbers are extrapolated from percentages given in the report.

volvement of the cervical spine and six, of the lumbar spine. Dahlin and Unni studied 418 patients who had monostotic fibrous dysplasia; only six patients had vertebral involvement. There have been six case reports of patients who had

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Case Report

A forty-one-year-old white man, who was otherwise healthy, had had pain in the leg for two weeks. A bone scan was made to rule out a stress fracture. The scan showed no abnormality of the lower extremity, but there was increased uptake in the second cervical vertebra.



FIG. 1

Plain lateral radiograph illustrating the lytic, expansile nature of the lesion of the second cervical vertebra. The typical ground-glass appearance of fibrous dysplasia is absent.

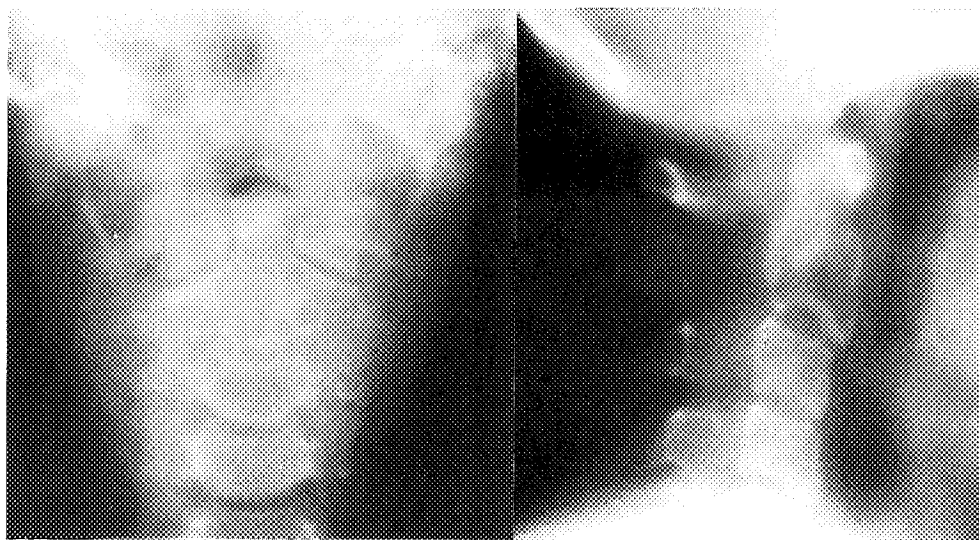


FIG. 2

Selected tomograms demonstrating destructive, expansile involvement of the entire vertebral body, odontoid process, and posterior elements. Note the disruption of the right facet joint of the second and third cervical vertebrae.

The patient's only symptom that was referable to the neck, which was elicited after the scan had been made, was a six-month history of occasional mild aching after he played volleyball. No numbness, weakness, or paresthesias of the extremities were noted. The medical and family histories were non-contributory. He had no soft-tissue lesions or *café-au-lait* spots. There was no local mass or tenderness, no limitation of motion of the neck, and no sensory or motor defects. The patient had normal reflexes and negative Babinski reflexes. The cranial nerves were intact, and sphincter tone was normal. Results of blood tests, including a routine

hemogram, and radiographs of the chest were also unremarkable.

The radiographs and tomograms showed an expansile, destructive lesion involving the anterior and posterior elements of the second cervical vertebra (Figs. 1 and 2). Additional testing included computed tomography and magnetic resonance imaging, which demonstrated slight narrowing of the spinal canal and no involvement of adjacent structures (Figs. 3 and 4).

Our differential diagnosis at that time included osteoblastoma, fibrous dysplasia, eosinophilic granuloma, aneurysmal bone cyst, and giant-cell tumor. Chordoma or a solitary metastasis were considered as remote possibilities.

Because the osseous involvement in the vertebra extended to the

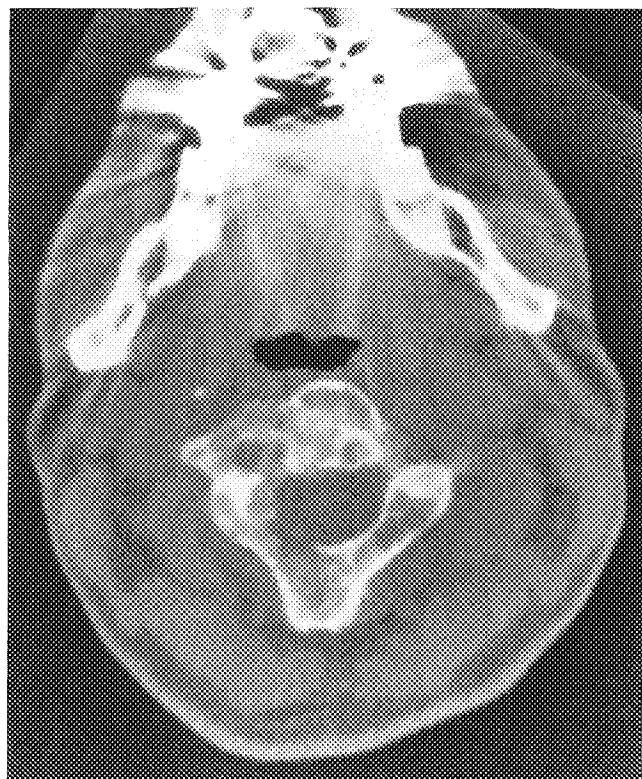


FIG. 3

Computed tomographic scan showing destruction of the anterior cortex of the right lamina and the anterior tubercle of the transverse process, as well as the absence of soft-tissue involvement or neural encroachment.

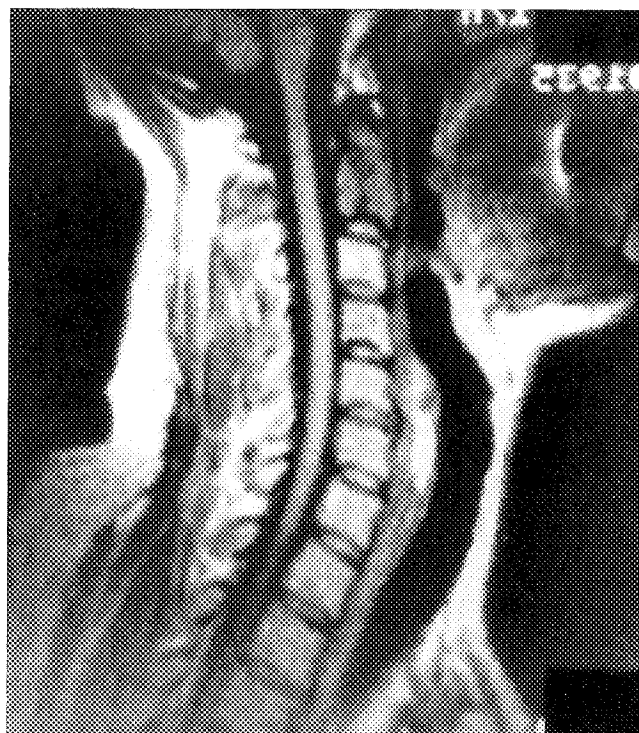


FIG. 4

The T2-weighted magnetic-resonance image demonstrates involvement of the entire vertebra, no encroachment on the spinal canal, and a possible longitudinal defect in the vertebral body.

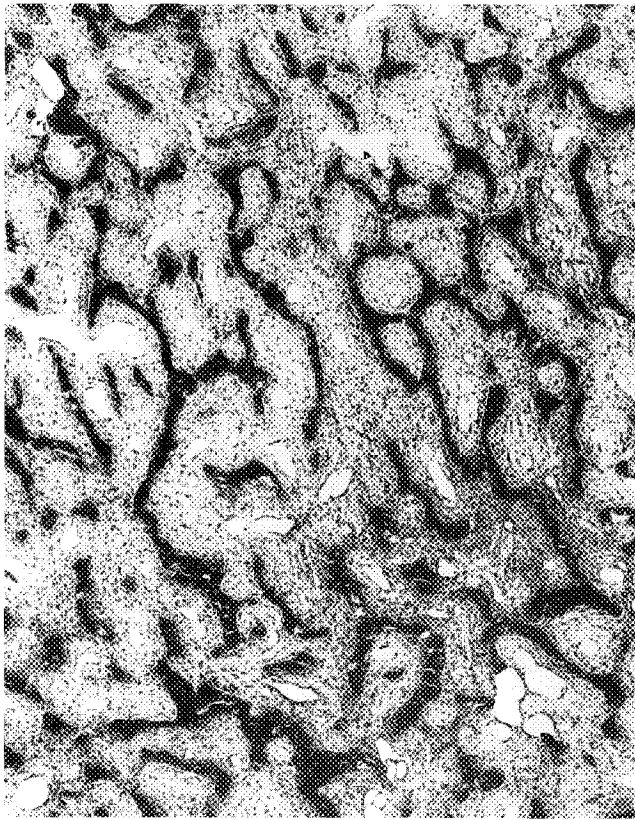


FIG. 5

The histological specimen shows fibrous dysplasia: slender, curved, misshapen bone, with interwoven fibrous tissue of moderate cellularity. This pattern often is referred to as having the appearance of alphabet soup (hematoxylin and eosin, $\times 200$).

posterior elements, a posterior approach was elected for the biopsy. This approach would enable us to do a posterior arthrodesis of the first, second, and third cervical vertebrae, if necessary, during the same operative session. We believed that, if the diagnosis based on the biopsy was favorable, stabilization would reduce the chance of pathological fracture. It also would afford greater safety if an anterior decompression was subsequently needed.

A halo was placed on the patient preoperatively, while he was awake. Routine posterior exposure was performed. The posterior elements of the second cervical vertebra were found to be porous, distorted, and very soft. The spinous process of the second cervical vertebra and a portion of the lamina were taken for histological examination. The spinal arthrodesis was performed with two grafts of corticocancellous bone from the posterior iliac crest; a sublaminar wire at the first cervical vertebra and two spinous-

process wires at the third cervical vertebra were used to stabilize the grafts. Additional cancellous bone was added on each side of the grafts.

The tissue of the lesion could be cut easily with a knife and was gritty and gray-yellow. On histological examination, the appearance was typical of fibrous dysplasia of bone (Fig. 5). Examination of a frozen section confirmed the diagnosis intraoperatively.

Osseous fusion was evident after fourteen weeks. Two and one-half years later, the patient had no pain, functional restriction, or neurological loss. There was no change in the radiographic appearance of the second cervical vertebra or the fusion mass.

Discussion

Because of the rarity of vertebral involvement by fibrous dysplasia, the preoperative diagnosis in this patient was difficult. The presence of a lesion was recognized only when a bone scan was made for an unrelated problem.

Of the diagnostic modalities that were used (except radiography), computed tomography provided the most information because it showed the osseous expansion and destruction. Whether treatment was needed is controversial. The patients reported on in the literature whose cases were similar were managed in various ways. When neurological findings were present, resection and arthrodesis were performed^{6,8}. Patients who had less severe symptoms or less extensive vertebral involvement have been managed conservatively^{7,14}. Russell and Chandler saw a patient who had polyostotic fibrous dysplasia, low-back pain, and a lesion of the anterosuperior aspect of the fifth lumbar vertebra. Spinal arthrodesis, the specifics of which were not reported, partially relieved the pain.

The case of our patient posed a problem not only of diagnosis but also of therapeutic planning. The symptoms that were referable to the neck were slight, and the radiographs did not provide diagnostic information. Biopsy was therefore necessary. Because the entire second cervical vertebra was affected, potentially compromising the stability of the neck in a particularly dangerous segment, arthrodesis seemed to be the logical option. When the histological examination revealed soft, disorganized bone that was susceptible to fracture or deformity, with possible disastrous neurological consequences, the decision to perform arthrodesis of the first, second, and third cervical vertebrae seemed reasonable.

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